



K1532
I

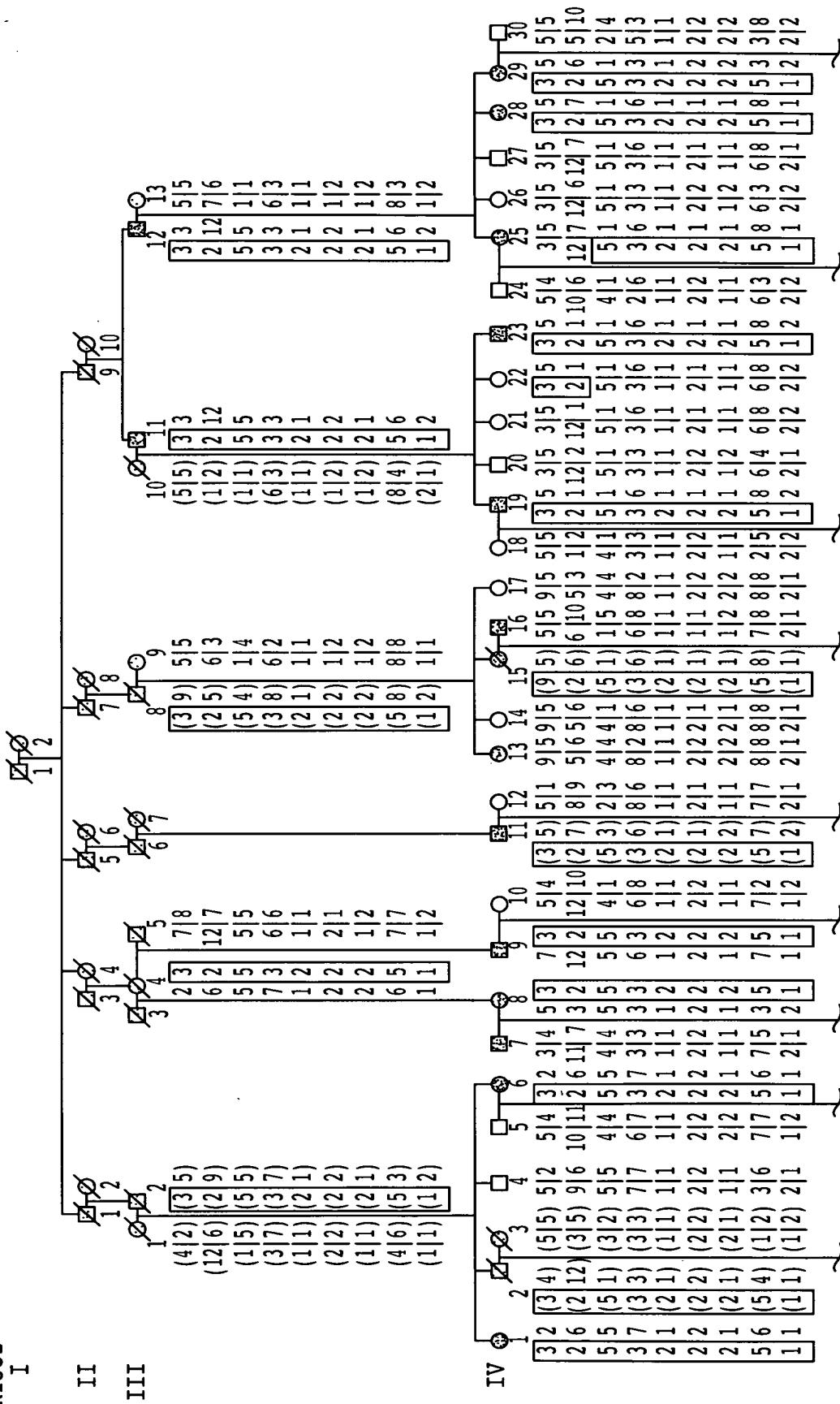


FIG. 1A



KVLQT1 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty Ref. 2323-151
SN 09/597,732, filed 19 June 2000

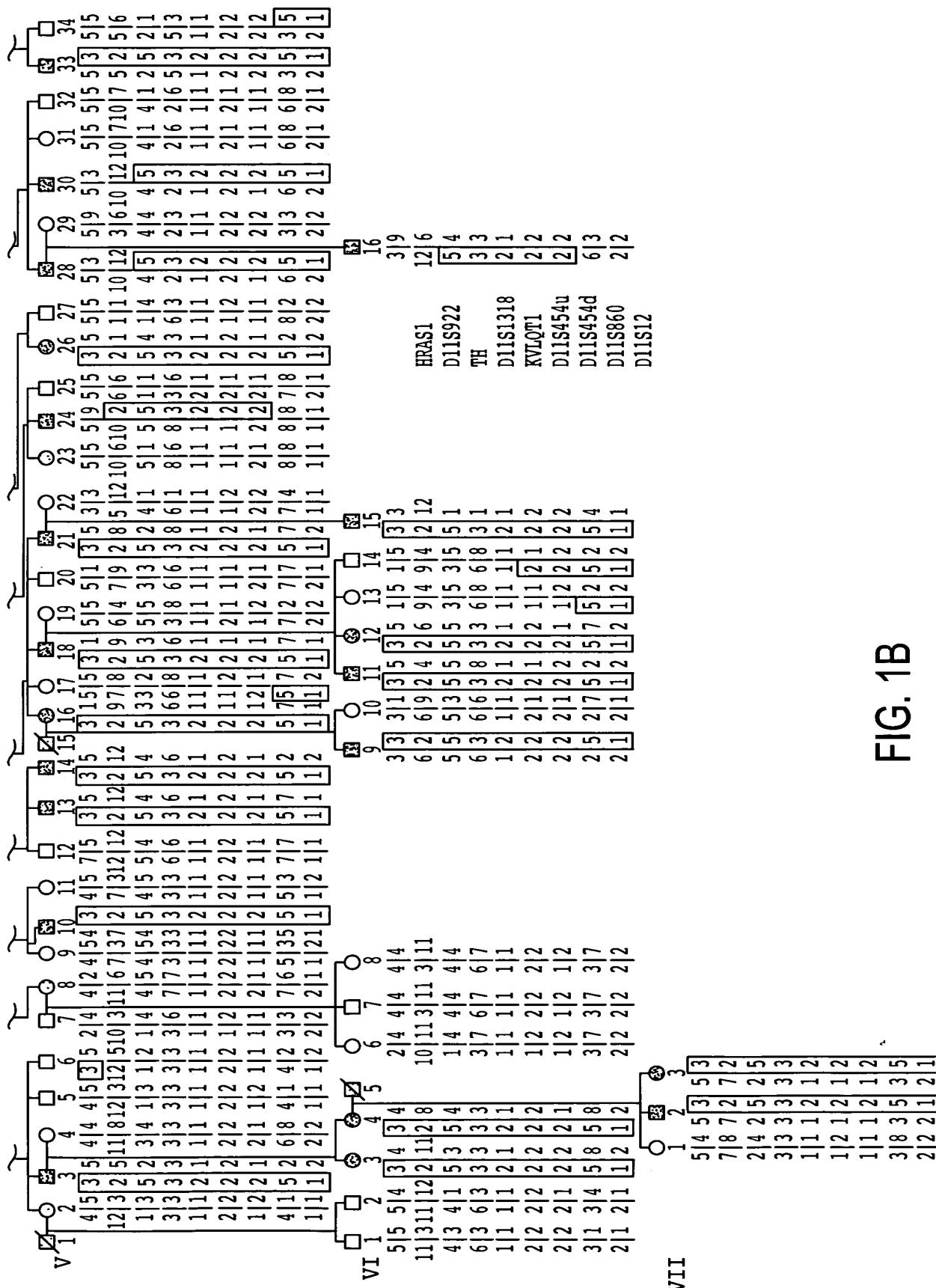


FIG. 1B



KVLQT1 - A Long QT Syndrome Gene
 Mark T. KEATING et al. - Atty. Ref. 2323-151
 SN 09/597,732, filed 19 June 2000

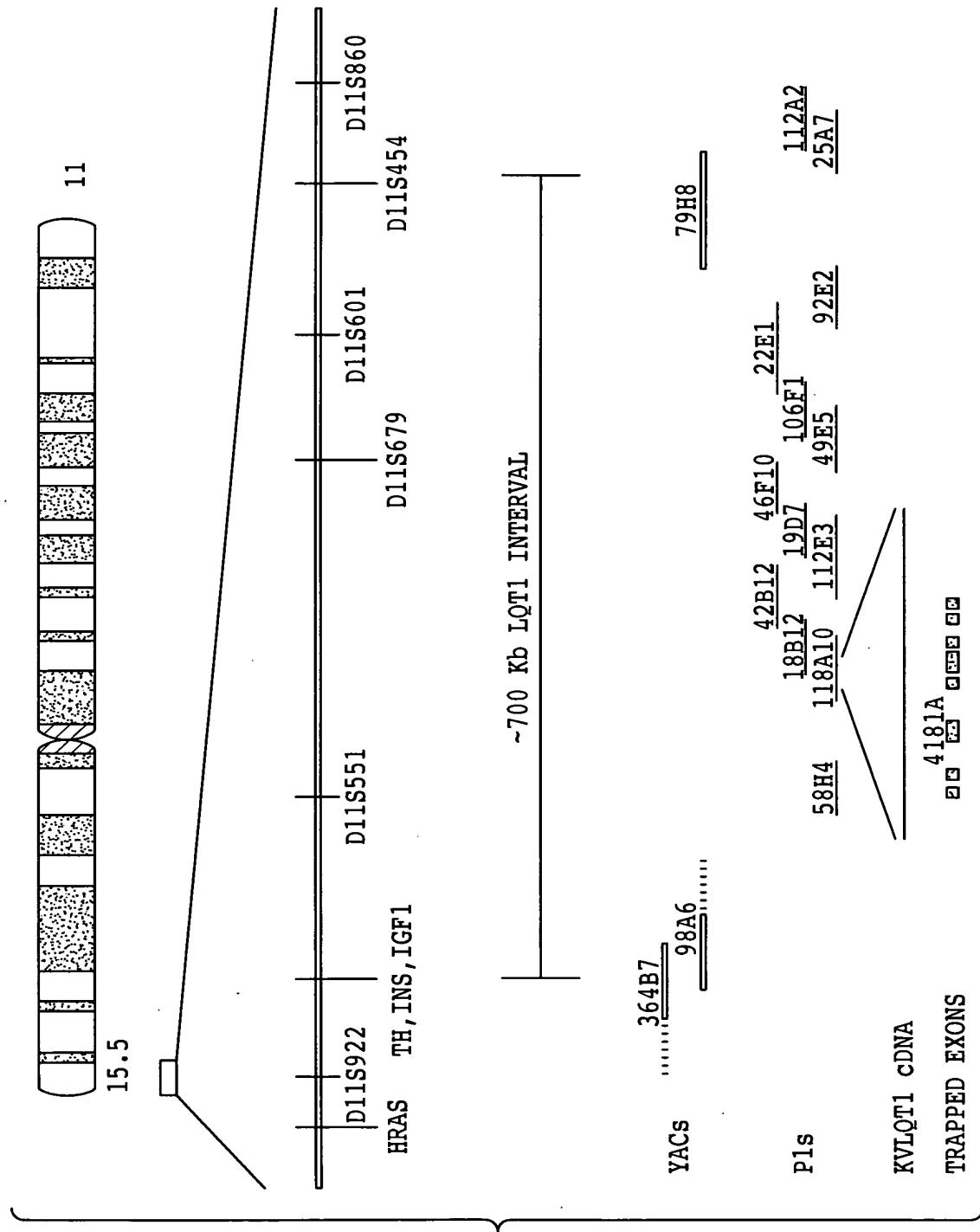


FIG. 2



KVLQT1 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty Ref. 2323-151
SN 09/597,732, filed 19 June 2000

8

101

2

5

FIG. 3

4



KVLQ11 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty Ref. 2323-151
SN 09/597,732, filed 19 June 2000

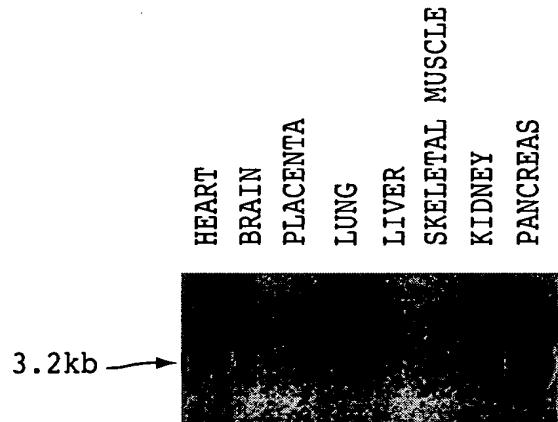


FIG. 4



CTGCCCCCTCCGGCCCCGCCCCGAGCGCCCGGGCTGGGCCGGCAGCGGCCCCCGCGGGGGCTGGCAGCAGTGGCTGCC-81
 CGCACTGCGCCGGGGCTCGCCTCGCTGCAGCTCCCGTGGAGCTGGCGAGGGCCTGGGGTGGGGCCCTGCCAGGGCCGGCGCAGC-243
 M A A A S S P P R A E R K R W G R L P G A R R G S -27
 GCGGGCCTGGCAAGAAGTGCCCCCTCGCTGGAGCTGGCGAGGGCCGGCGCGCTACGGCCCAC-324
 A G L A K K C P F S L E L A E G G P A G G A L Y A P I -54
 GCGCCCGGGCGCCCGAGGTCCCGCGCCCCCTCGCTCCCCGGCCGCGCCCGCGCCCCAGTTGCCCTCCGACCTTGGCCCC-405
 A P G A P G P A P P A S P A A P A A P P V A S D L G P -81
 CGGCCGCCGGTGAGCCTAGACCCGGCGCTCCATCTACAGCACGCCGCCGGTGTGGCGCGCACCCACGTCCAGGGC-486
 R P P V S L D P R V S I Y S T R R P V L A R T H V Q G -108

CGCGTCTACAACCTCCTCGAGCGTCCCACCGGCTGGAAATGCTTCGTTACCACTTCGCCGTCTCCTCATCGTCTGGTC-567
 R V Y N F L E R P T G W K C F V Y H F A V F L I V L V -135

▼ S1

TGCTCATCTCAGCGTGCTGTCCACCATCGAGCAGTATGCCGCCCTGGCCACGGGACTCTCTCTGGATGGAGATCGTG-648
 C L I F S V L S T I E Q Y A A L A T G T L F W M E I V -162

▼ S2

CTGGTGGTGTCTCGGGACGGAGTACGTGGTCCGCCCTGGTCCGCCCTGGATGGCCAGCAAGTACGTGGCCTCTGGCTGGC-729
 L V V F F G T E Y V V R L W S A G C R S K Y V G L W G -189

CGCCTGCCCTTGCCCCGAAGCCCATTCCATCATCGACCTCATCGTGGCTGGCCCTCATGGTGGCTCTGGCTGGC-810
 R L R F A R K P I S I I D L I V V V A S M V V L C V G -216

▼ S3

TCCAAGGGCAGGTGTTGCCACGTCGCCATCAGGGGCATCCGCTTCGCAGATCCTGAGGATGCTACACGTCGACCGC-891
 S K G Q V F A T S A I R G I R F L O I L R M L H V D R -243

▼ S4

CAGGGAGGCACCTGGAGGCTCTGGCTCCGTGGCTTCATCCACCGCCAGGAGCTGATAACCACCTGTACATGGCTTC-972
 Q G G T W R L L G S V V F I H R Q E L I T T L Y I G F -270

CTGGGCCTCATCTCCTCGTACTTGTGTACCTGGCTGAGAAGGACGGTGAACGAGTCAGGCCGCGTGGAGTTGGC-1053
 L G L I F S S Y F V Y L A E K D A V N E S G R V E F G -297

▼ S5

AGCTACGCAGATGCGCTGTGGTGGGGGGTGGTCACAGTCACCCATCGCTATGGGACAAGGTGCCAGACGTGGTC-1134
 S Y A D A L W W G V V T V T I G Y G D K V P Q T W V -324

Pore

GGGAAGACCATGCCCTCTGCTCTGTCTTGCCATCTCCTCTGGCTCCAGGGGGATTCTGGCTGGGGTTT-1215
 G K T I A S C F S V F A I S F F A L P A G I L G S G F -351

▼ S6

GCCCTGAAGGTGCAGCAGAAGCAGAGGCAGAACGACTTCAACCGCAGATCCGGCGGGCAGCCTCACTCATTAGACCGCA-1296
 A L K V Q Q K Q R Q K H F N R Q I P A A A S L I Q T A -378

TGGAGGTGCTATGCTGCCGAGAACCCGACTCCTCACCTGGAAAGATCTACATCCGAAGGCCCCCGGAGCCACACTCTG-1377
 W R C Y A A E N P D S S T W K I Y I R K A P R S H T L -405

CTGTCACCCAGCCCCAACCAAGAAGTCTGGTGGTAAAGAAAAAAAGTTCAAGCTGGACAAAGACAATGGGTGACT-1458
 L S P S P K P K K S V V V K K K F K L D K D N G V T -432

CCTGGAGAGAAGATGCTCACAGTCCCCATATCACGTGCGACCCCCAGAAGAGCGGGCTGGACCACTCTGTGAC-1539
 P G E K M L T V P H I T C D P P E E R R L D H F S V D -459

GGCTATGACAGTTCTGTAAGGAAGAGCCCAACACTGCTGGAAAGTGAGCATGGCCATTCTAGAGAACCAACAGCTTCGCC-1620
 G Y D S S V R K S P T L L E V S M P H F M R T N S F A -486

GAGGACCTGGACCTGGAGGGGAGACTCTGCTGACACCCATCACAGCTGGGGAACACCACCGGAC-1701
 E D L D L E G E T L L T P I T H I S Q L R E H H R A T -513

ATTAAGGTCAATTGACGCATGCAGTACTTGTGGCAAGAAGAAATTCCAGCAAGCGCGGAAGCCTTACGATGTGGGGAC-1782
 I K V I R R M Q Y F V A K K K F Q Q A R K P Y D V R D -540

GTCATTGAGCAGTACTCGCAGGGCACCTCAACCTCATGGTGGCATCAAGGAGCTGCAGAGGAGGCTGGACCAAGTCCATT-1863
 V I E Q Y S Q G H L N L M V R I K E L Q R R L D Q S I -567

FIG. 5A



KVLQT1 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty. Ref. 2323-151
SN 09/597,732, filed 19 June 2000

▼
GGGAAGCCCTCACTGTTCATCTCCGTCTCAGAAAAGAGCAAGGATCGCGCAGCAACACGATCGGCGCCCTGAACCGA-1944
G K P S L F I S V S E K S K D R G S N T I G A R L N R -594
▼
GTTAGAAGACAAGGTGACCGCAGCTGGACCAGAGGCTGGCACTCATCACCGACATGCTTACCCAGCTGCTCTCCCTGACGGT-2025
V E D K V T Q L D Q R L A L I T D M L H Q L L S L H G -621
GGCAGCACCCCCGGCAGCGCGGGCCCCCAGAGAGGGGGGCCACATCACCCAGCCCTGGGCAGTGGCGCTCCGTC-2106
G S T P G S G G P P R E G G A H I T Q P C G S G G S V -648
GACCTGAGCTCTTCTGCCCCAGAACACCCCTGCCCACCTACGAGCAGCTGACCGTGGCCAGGAGGGGCCGATGAGGGG-2187
D P E L F L P S N T L P T Y E Q L T V P R R G P D E G -675
TCCTGAGGAGGGGATGGGGCTGGGGATGGGCTGAGTGAGGAGGGAGGCAAGAGTGGCCCACTGGCCCTCTGAAG-2268
S * -676
GAGGCCACCTCTAAAAGGCCAGAGAGAACAGGCCACTCTCAGAGGCCAATACCCATGGACCATGCTGTCTGGCAC-2349
AGCCTGCACTTGGGGCTCAGCAAGGCCACCTCTCCTGGCCGGTGTGGGGGCCGTCAGGTCTGAGTTGTTACCCA-2430
AGCGCCCTGGCCCCACATGGTGTGATGTTGACATCACTGGCATGGTGGTGGGACCCAGTGGCAGGGCACAGGGCCTGGCCC-2511
ATGTATGCCAGGAAGTAGCACAGGCTGAGTGCAGGCCACCCCTGCCAGGCACAGGCAGGGCAGGACCCAGCCTGACTACAGGGCAC-2592
ACCCCTGGACCCAGCCTAAATCCAGGACCCCTGCCAGGCACAGGCAGGGCAGGACCCAGCCTGACTACAGGGCAC-2673
CGGCAATAAAAGCCCAGGAGGCCATTGGAGGGCTGGCTGGCTCCACTCTCAGGAATGCTGACCATGGGCAGG-2754
AGACTGTGGAGACTGCTCTGAGCCCCACTTCCCTGGGTAGACTGCCAGCTCTCTAGCTGGAGGGACCCCTGCCCTCTCCGCCCCGAGTGGTTGAGT-2835
GGGGGGAAACGCCCACTTCCCTGGGTAGACTGCCAGCTCTCTAGCTGGAGGGACCCCTGCCCTCTCCGCCCCGAGTGGTTGAGT-2916
ACTGTGCGTGGGGCTCCCGCCTCCAACCCCTGCCAGTCCCAGCAGGCCAGCCAACACACAGAAGGGGACTGCCACCTCC-2997
CCTTGCCAGCTGCTGAGCCAGAGAAAGTACAGGTTCTACACAGGACAGGGGTTCTCTGGCATTACATGCATAGAA-3078
ATCAATAATTGTGGTATTGGATCTGTGTTAATGAGTTCACAGTGTGATTATTAAATTGTGCAAGCTTTC-3159
CTAATAAACGTGGAGAACAC(A)n -3180

FIG. 5B



KVLQT1 - A Long QT Syndrome Gene
 Mark T. KEATING et al. - Atty Ref. 2323-151
 SN 09/597,732, filed 19 June 2000

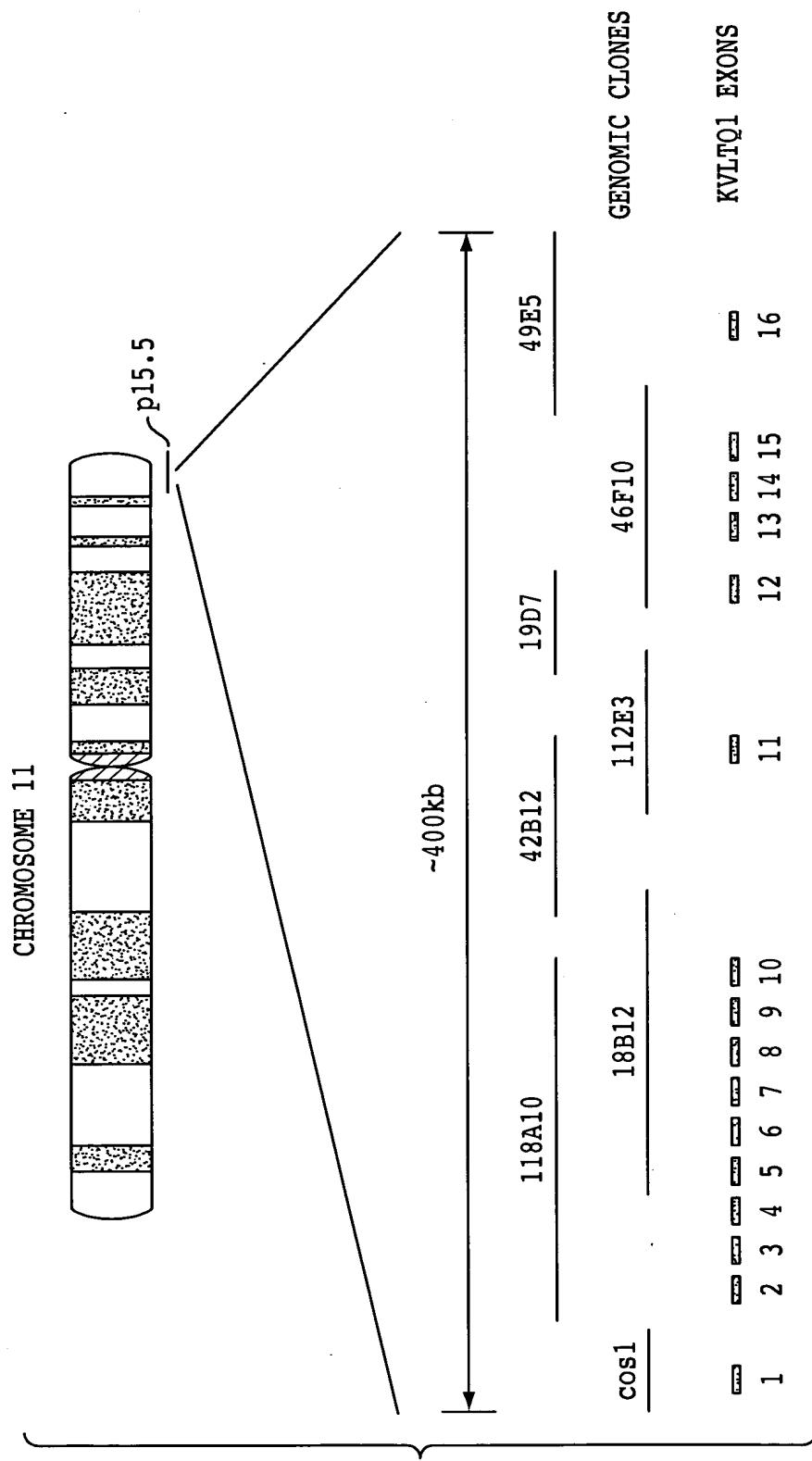


FIG. 6

8



KVLQT1 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty. Ref. 2323-151
SN 09/597,732, filed 19 June 2000

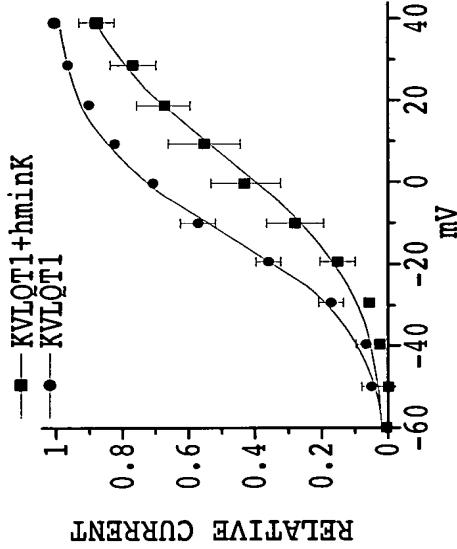


FIG. 7B

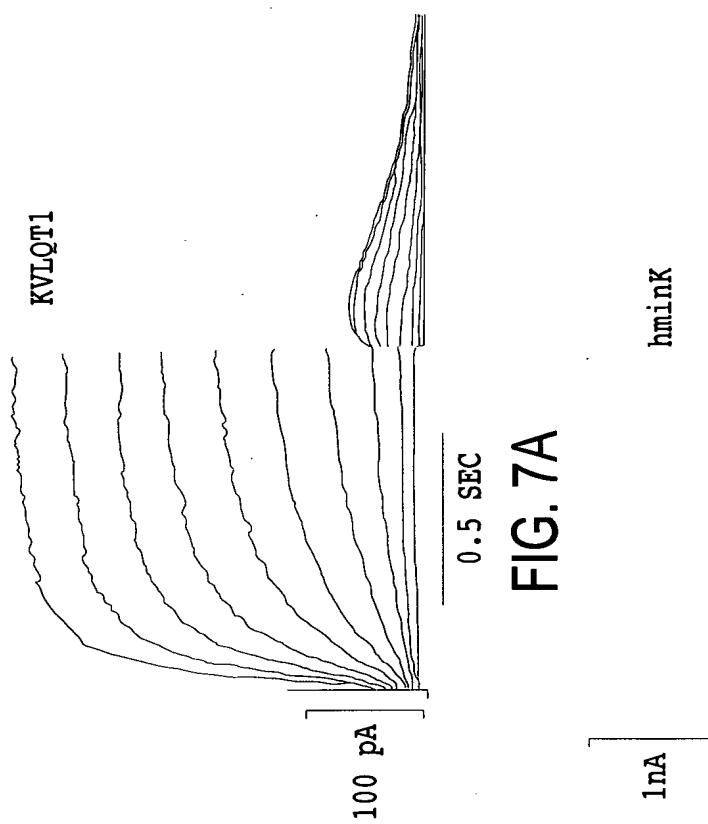


FIG. 7A

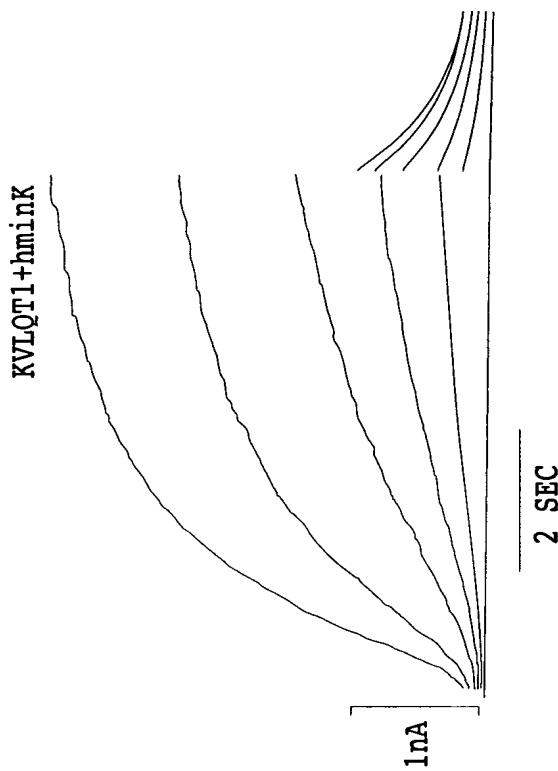


FIG. 7E

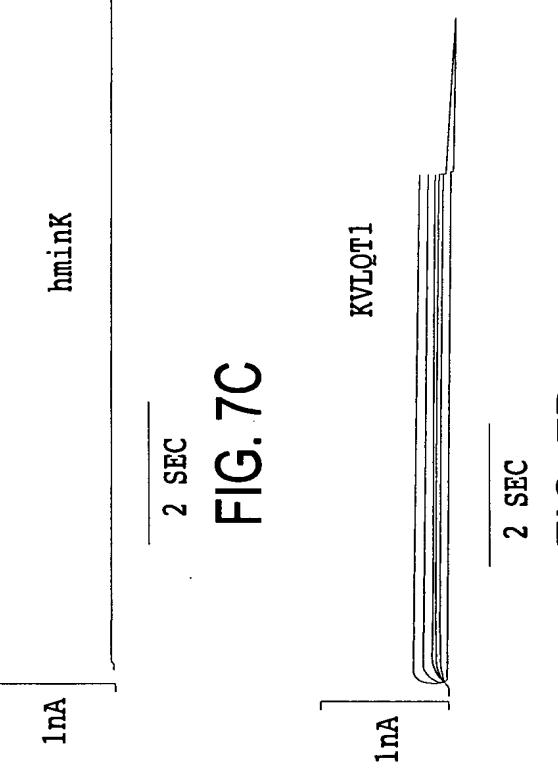


FIG. 7D



KVLQT1 - A Long QT Syndrome Gene
 Mark T. KEATING et al. - Atty Ref. 2323-151
 SN 09/597,732, filed 19 June 2000

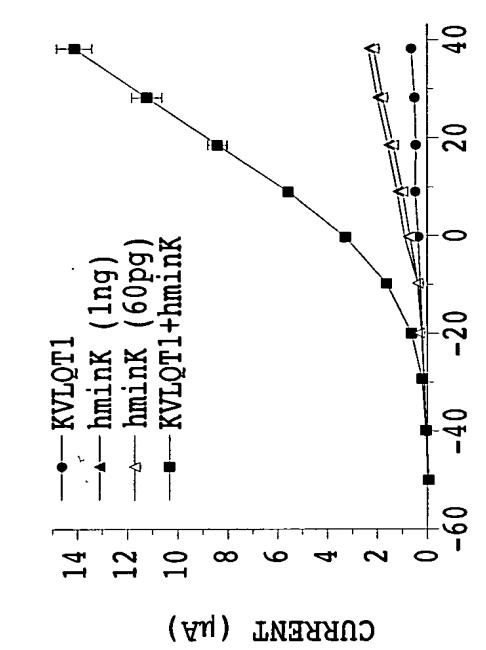


FIG. 9A

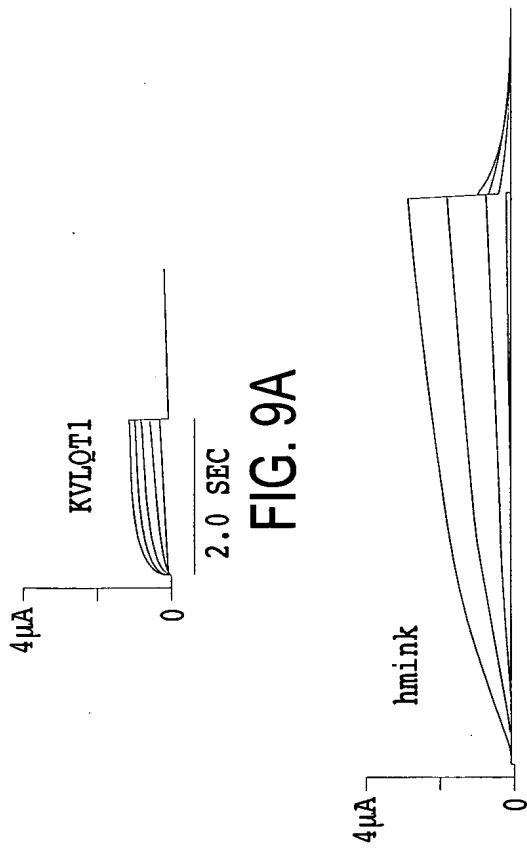


FIG. 9B

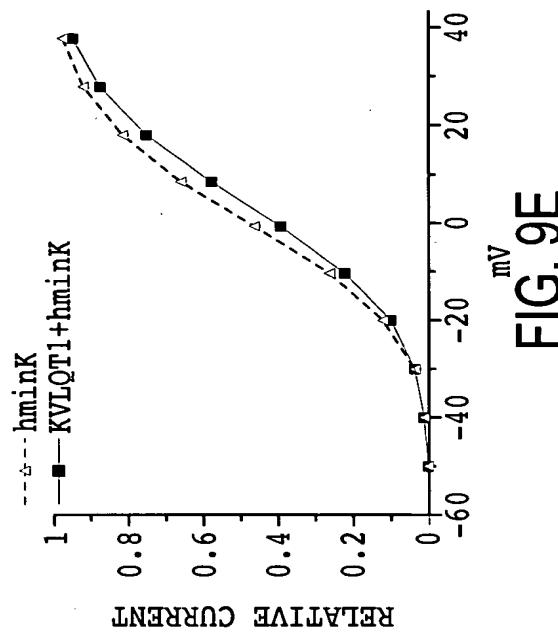


FIG. 9C

FIG. 9D

FIG. 9E

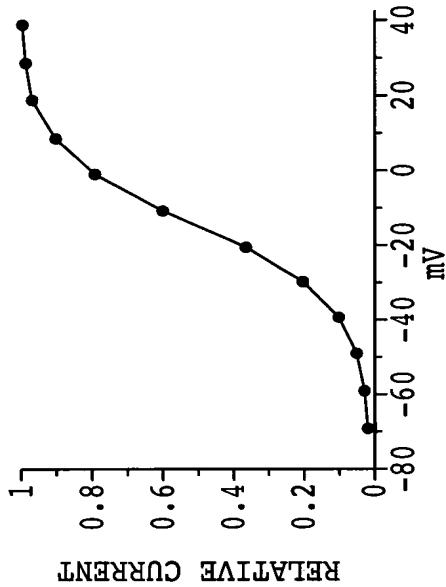


FIG. 8B

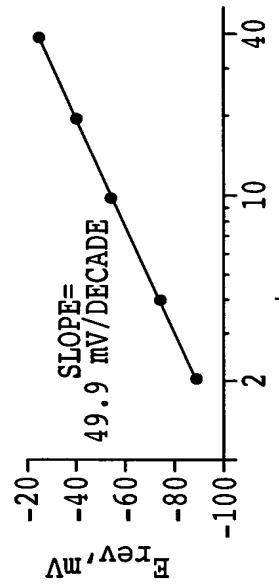


FIG. 8C

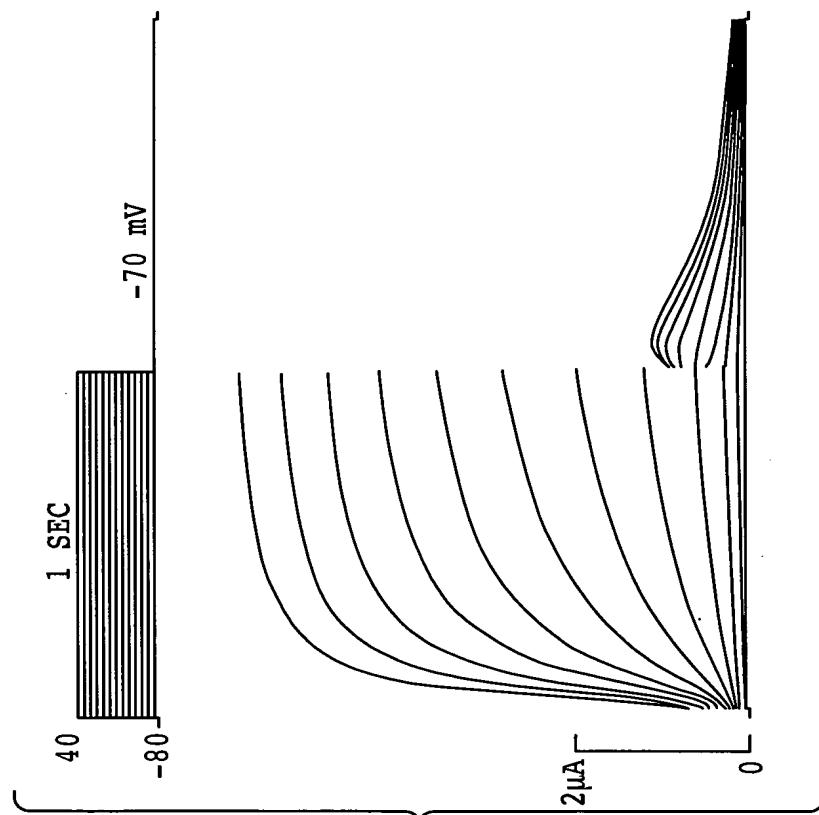


FIG. 8A



KVLQT1 - A Long QT Syndrome Gene
 Mark T. KEATING et al. - Atty. Ref. 2323-151
 SN 09/597,732, filed 19 June 2000

XENOPUS

MNENA
AINS
LYEAIPLP
QDGSS
NGQRQEDRQ
ANSFELK
RETLVATD
PPRPT

HUMAN

QGRVYNFLERPTG
WKCFVYHFAVFLIVL
|||||

XENOPUS

INLDPRVSIYSGRRPLFSRTNIQGRVYNFLERPTG
WKCFVYHFTVFLIVL

HUMAN

S1 VCLIFSVLSTIEQYAALATGTLFWMEIVLVVFFGTEYV
VRLWSAGCRSKY
|||||

XENOPUS

ICLIFSVLSTIQQYNNLATETLF
FWMEIVLVVFFGA
YV
VRLWSAGCRSKY

HUMAN

VGLWGRRLRFARKP
ISIIDLIVV
VASMV
LCVGS
KGQV
FATSAIRGIRFLQ
|||||

XENOPUS

VGWGRRLRFARKP
ISVIDLIVV
VASV
VLCVGS
NGQV
FATSAIRGIRFLQ
A

HUMAN

ILRMLHVDRQGGT
WRL
LGSVV
FIHRQEL
ITTL
YIGFLGLI
FSSYFV
LAE
|||||

XENOPUS

ILRMLHVDRQGGT
WRL
LGSVV
FIHRQEL
ITTL
YIGFLGLI
FSSYFV
LAE

HUMAN

KDAVN
ESGRVEFG
SYADAL
WWGV
VT
TTIGY
GDKV
PQT
WVGK
TIA
SCF
SV
|||

Pore

XENOPUS

KDAIDSS
GEYQFG
SYADAL
WWGV
VT
TTIGY
GDKV
PQT
WIGK
TIA
SCF
SV

HUMAN

FAISFF
FALP
AGIL
GSGF
ALKV
QQK
Q
RQK
HF
NRQ
I
P
AA
AS
LI
Q
TAW
RCY
AA
|||||

XENOPUS

FAISFF
FALP
AGIL
GSGF
ALKV
QQK
Q
RQK
HF
NRQ
I
P
AA
AS
LI
Q
TAW
RCY
AA

HUMAN

ENPDS
STWK
IYIRK
APRS
HTLL
SPSP
PKK
SVV
KKK
F
L
D
N
G
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|||

XENOPUS

ENPDS
ATWK
IYIRK
QSRN
HHIM
SPSP

HUMAN

EKMLTV
PHITCD
PPEERR
LDHF
SVDGY
DSS
VRKS
PTL
LEVS
MPH
FMRT
NS

HUMAN

FAEDLD
LEGET
LLTP
I
THIS
QLRE
H
RATI
K
VIR
RMQY
F
VAK
KKF
QQARK

HUMAN

PYD
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IEQ
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SQG
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HUMAN

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HUMAN

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R
R
G
P
D
E
G
S

FIG. 10

12

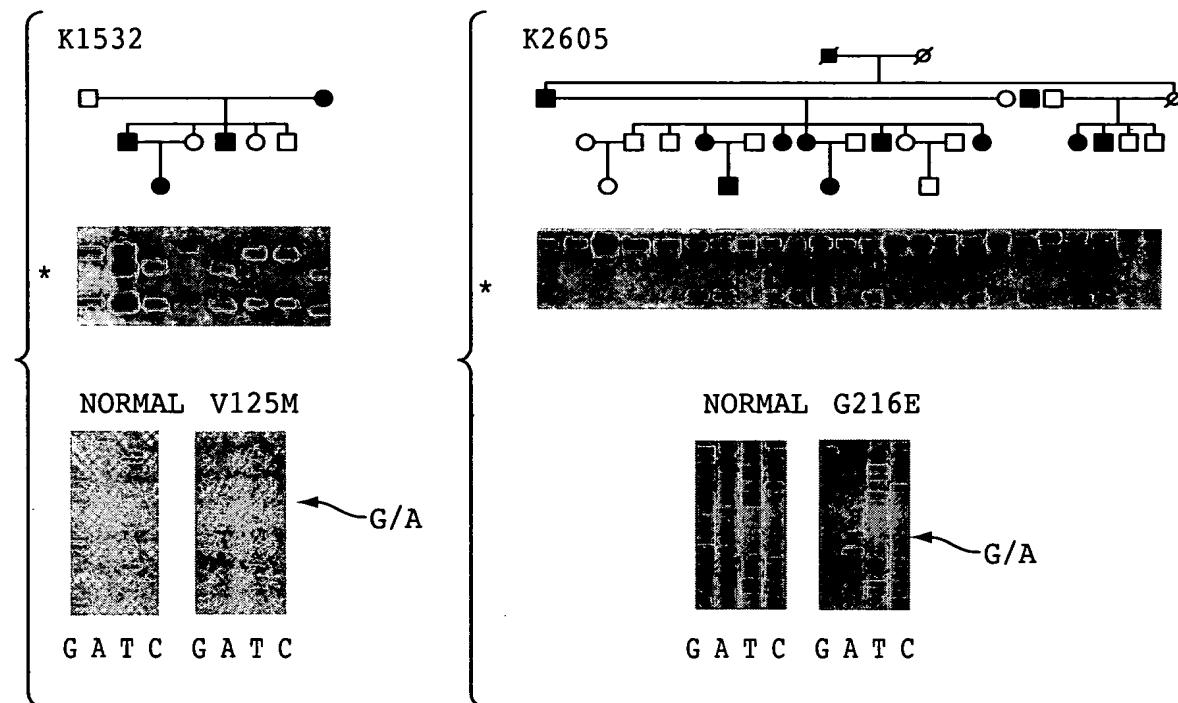


FIG. 11A

FIG. 11B

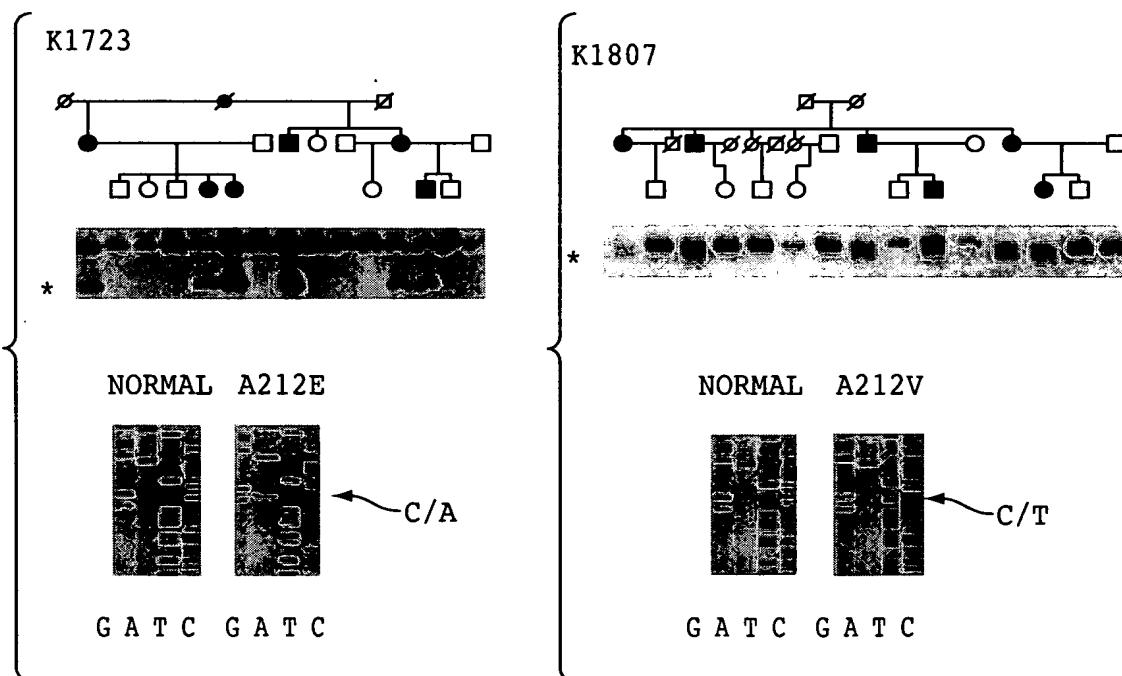


FIG. 11C

FIG. 11D



KVLQT1 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty. Ref. 2323-151
SN 09/597,732, filed 19 June 2000

FIG. 12A

FIG. 12B

FIG. 12C

FIG. 12D



KVLQT1 - A Long QT Syndrome Gene
 Mark T. KEATING et al. - Atty Ref. 2323-151
 SN 09/597,732, filed 19 June 2000

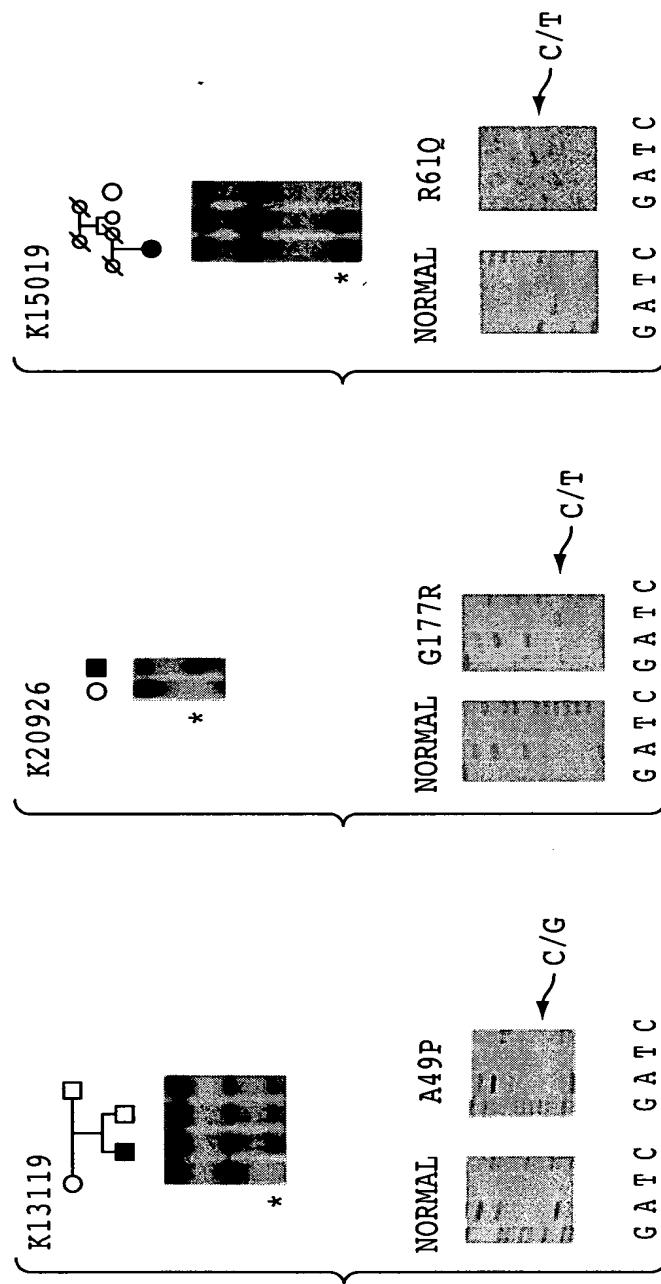


FIG. 12G

FIG. 12F

FIG. 12E



KVLQT1 - A Long QT Syndrome Gene
 Mark T. KEATING et al. - Atty. Ref. 2323-151
 SN 09/597,732, filed 19 June 2000

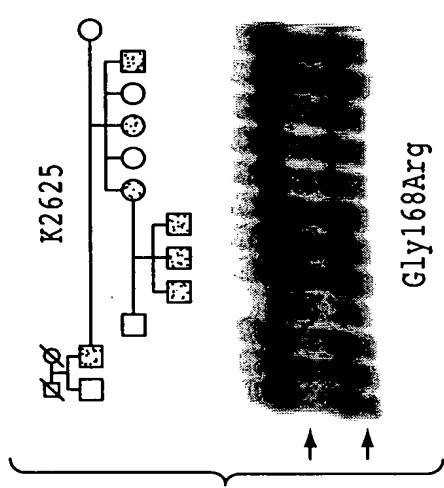


FIG. 12H
 Gly168Arg

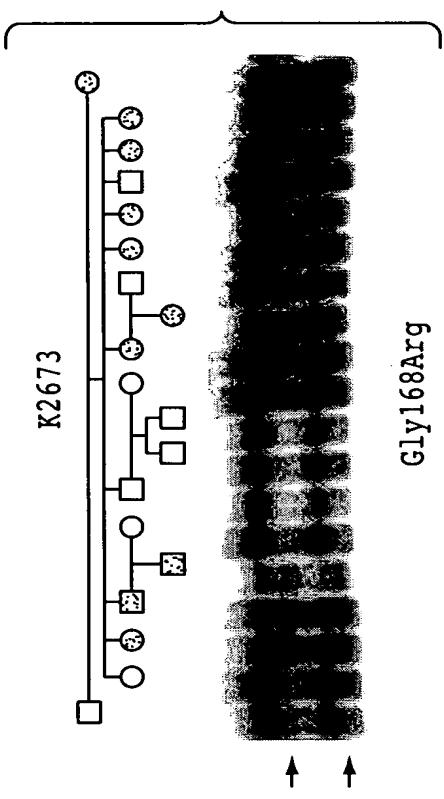


FIG. 12I
 Gly168Arg

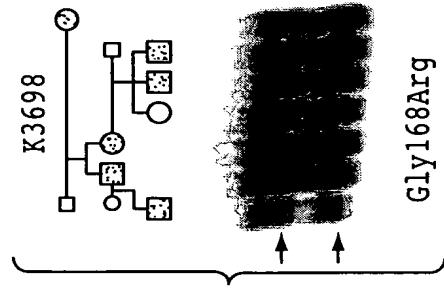


FIG. 12J
 Gly168Arg

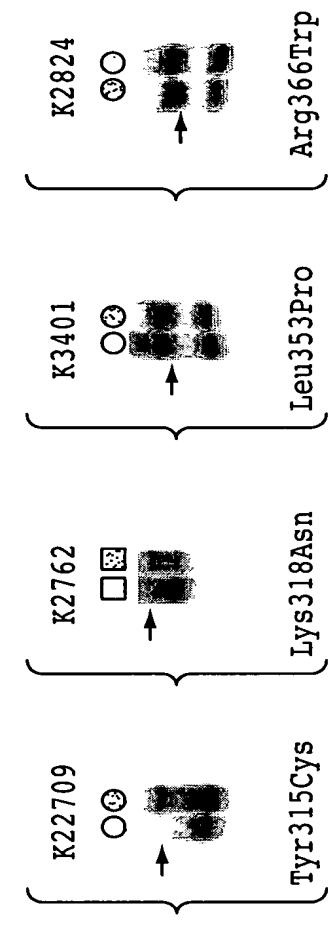


FIG. 12K
 Gly314Ser

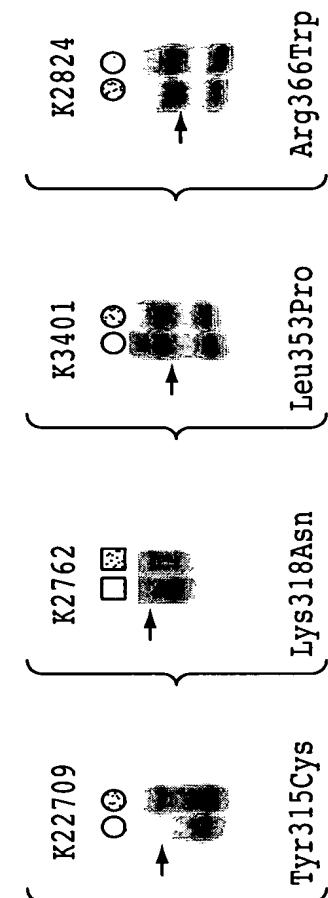


FIG. 12L
 Lys318Asn

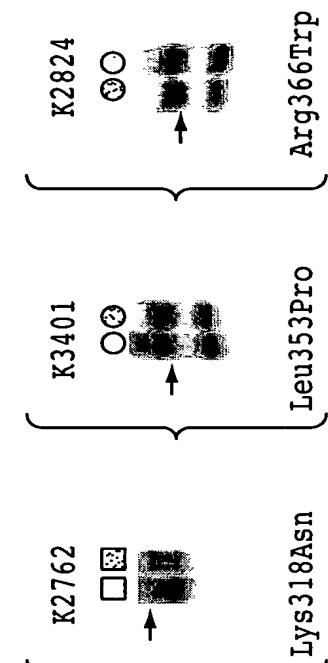


FIG. 12M
 Leu353Pro

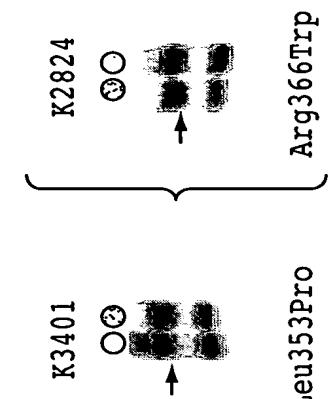


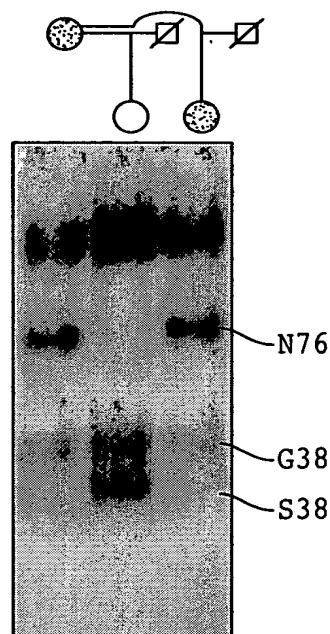
FIG. 12N
 Arg366Trp

FIG. 12O



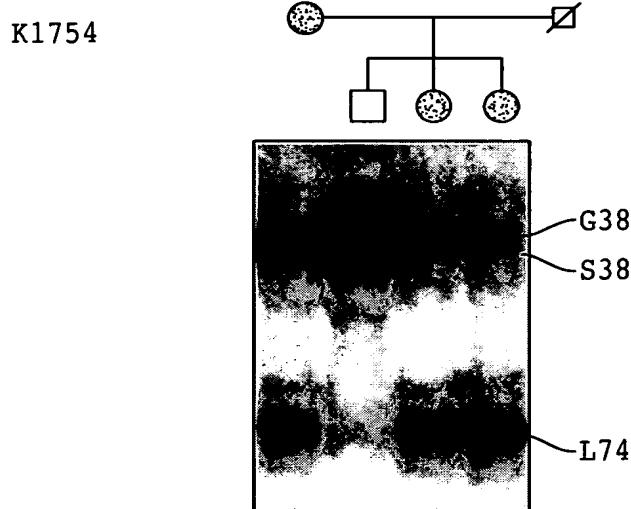
KVLQT1 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty Ref. 2323-151
SN 09/597,732, filed 19 June 2000

K1789



NORMAL	CAC	TCG	AAC	GAC	CCA	TTC	AAC
	H	S	N	D	P	F	N
MUTANT	CAC	TCG	AAC	AAC	CCA	TTC	AAC
	H	S	N	N	P	F	N

FIG. 13A



NORMAL	CTG	GAG	CAC	TCG	AAC	GAC	CCA
	L	E	H	S	N	D	P
MUTANT	CTG	GAG	CAC	TTG	AAC	GAC	CCA
	L	E	H	L	N	D	P

FIG. 13B

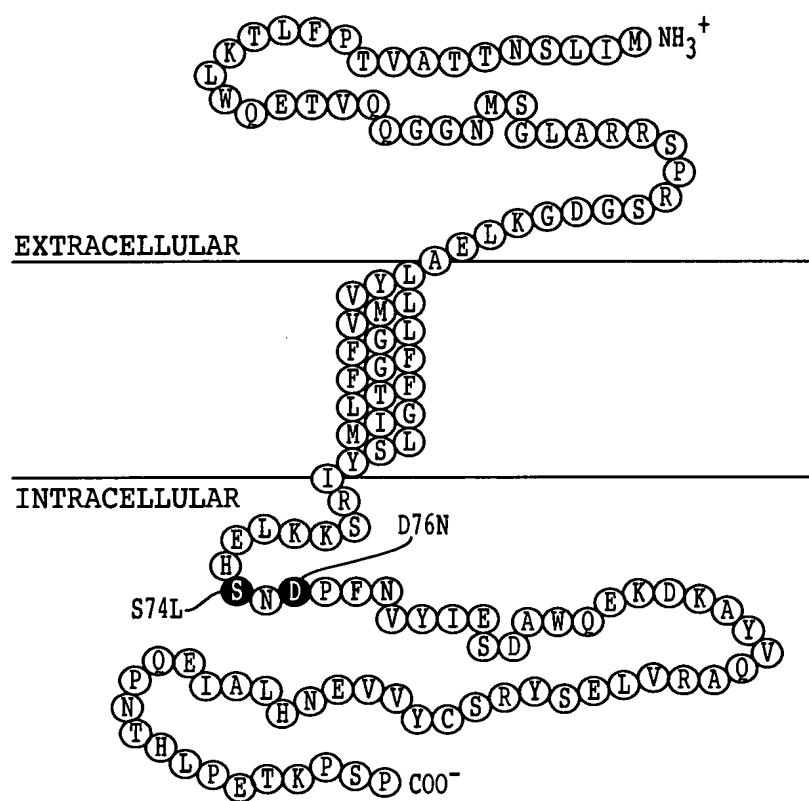
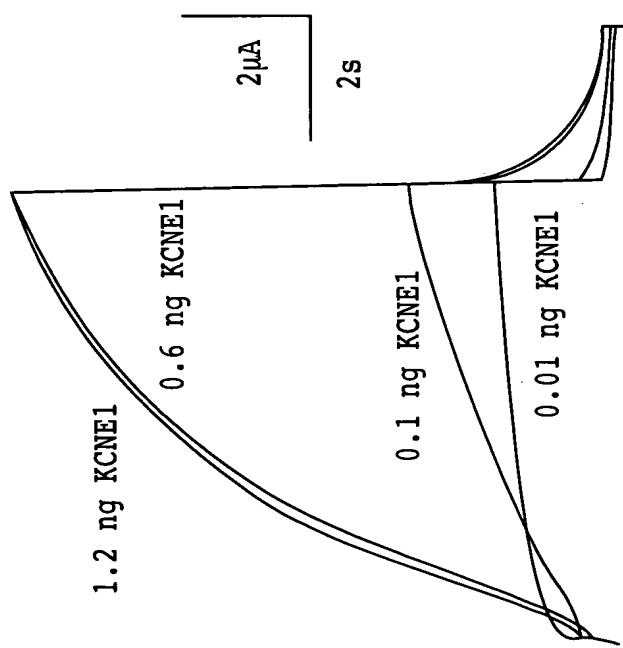
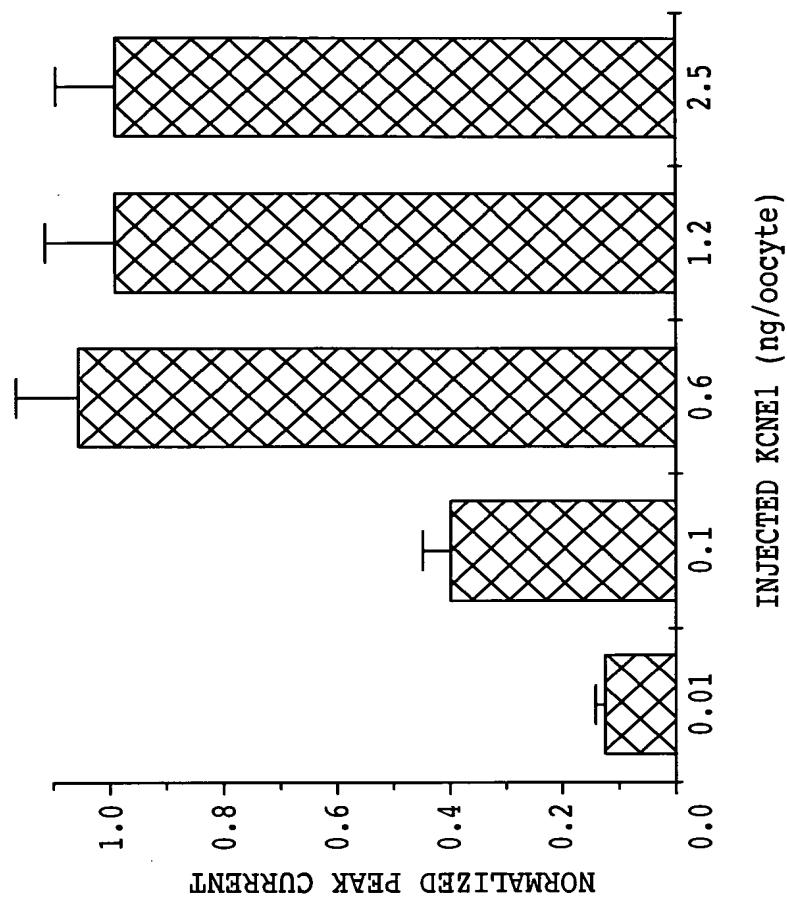


FIG. 13C



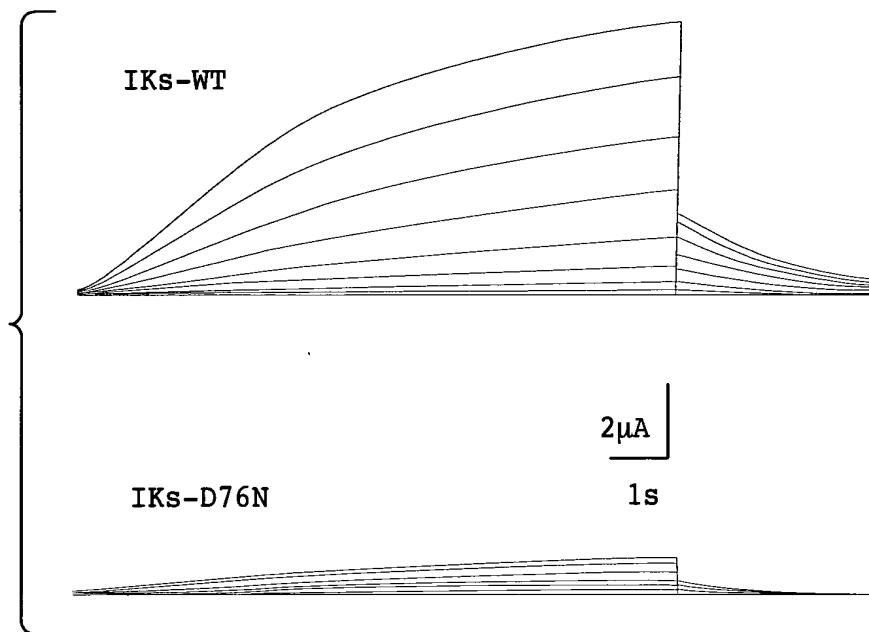


FIG. 15A

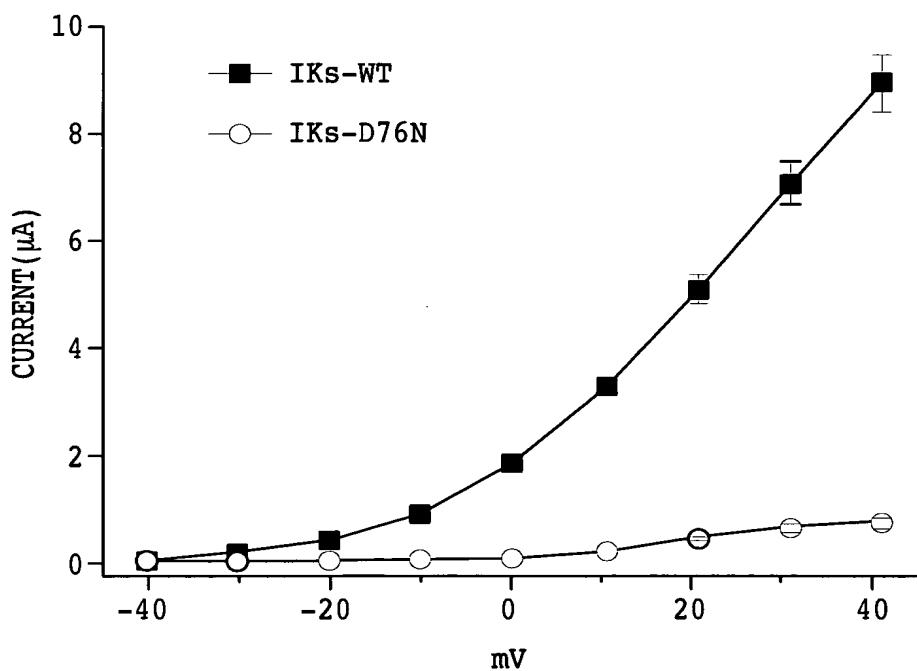


FIG. 15B



KVLQT1 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty Ref. 2323-151
SN 09/597,732, filed 19 June 2000

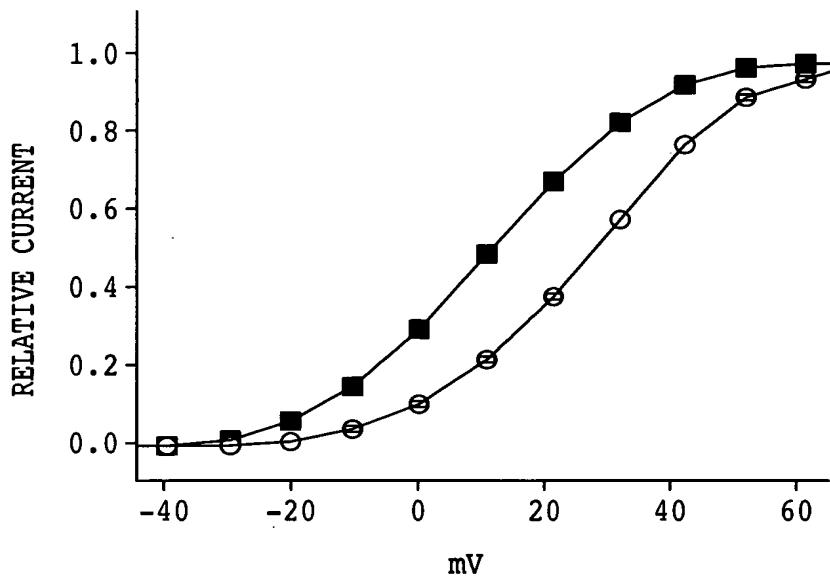


FIG. 15C

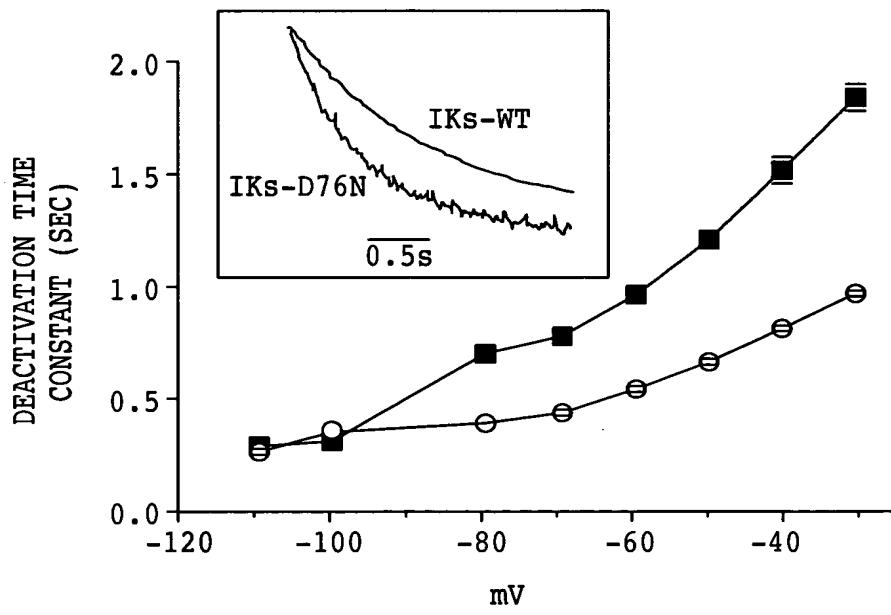


FIG. 15D

22



KVLQT1 - A Long QT Syndrome Gene
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SN 09/597,732, filed 19 June 2000

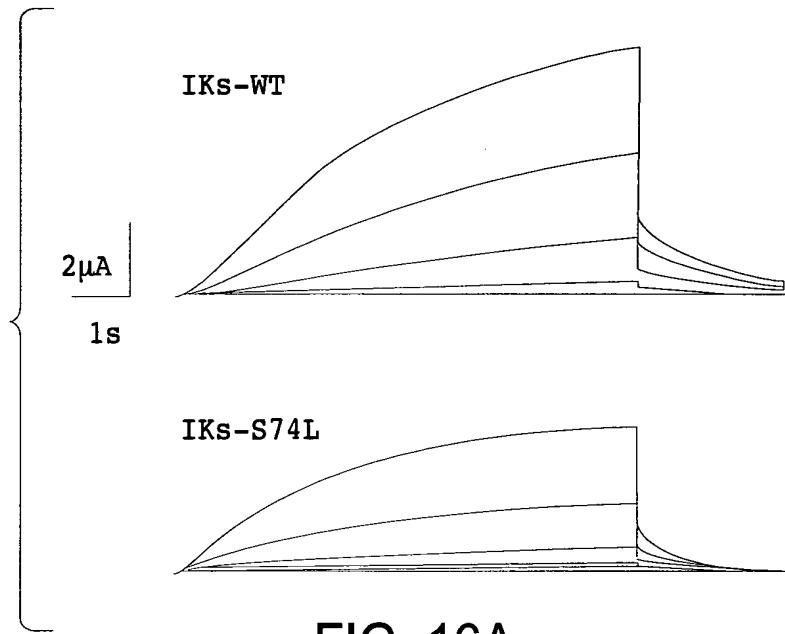


FIG. 16A

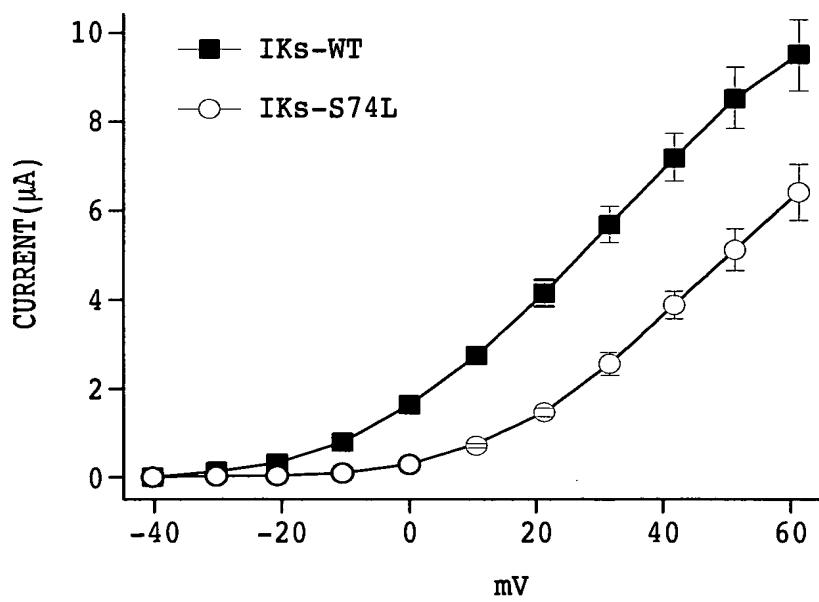


FIG. 16B



KVLQT1 - A Long QT Syndrome Gene
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SN 09/597,732, filed 19 June 2000

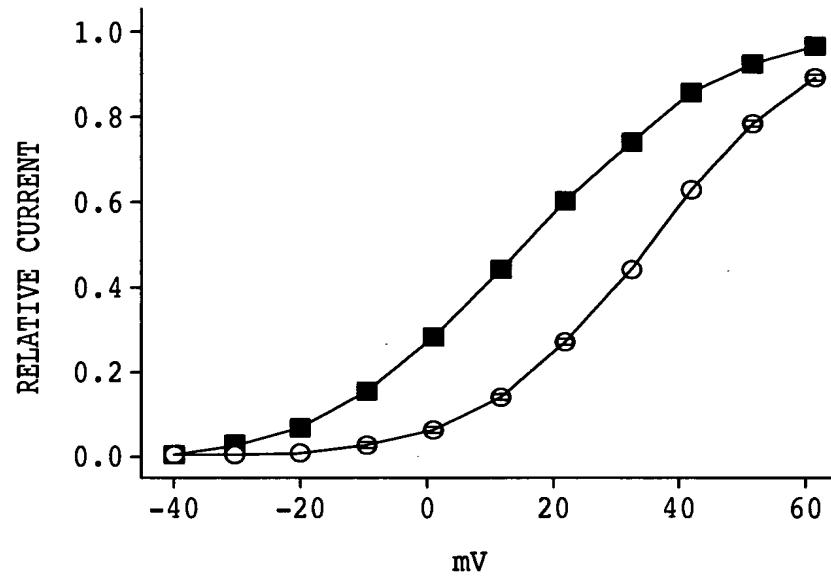


FIG. 16C

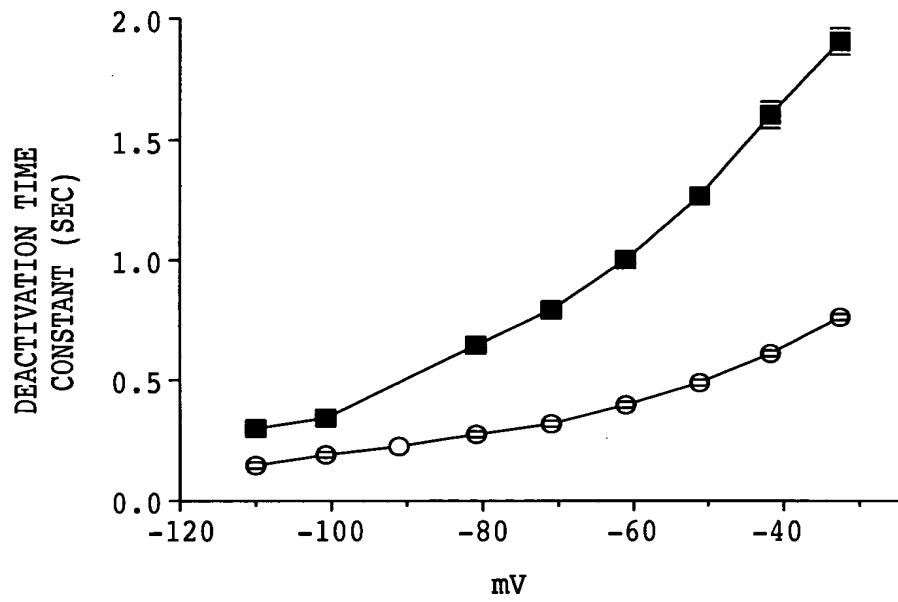


FIG. 16D

24



KVLQT1 - A Long QT Syndrome Gene
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SN 09/597,732, filed 19 June 2000

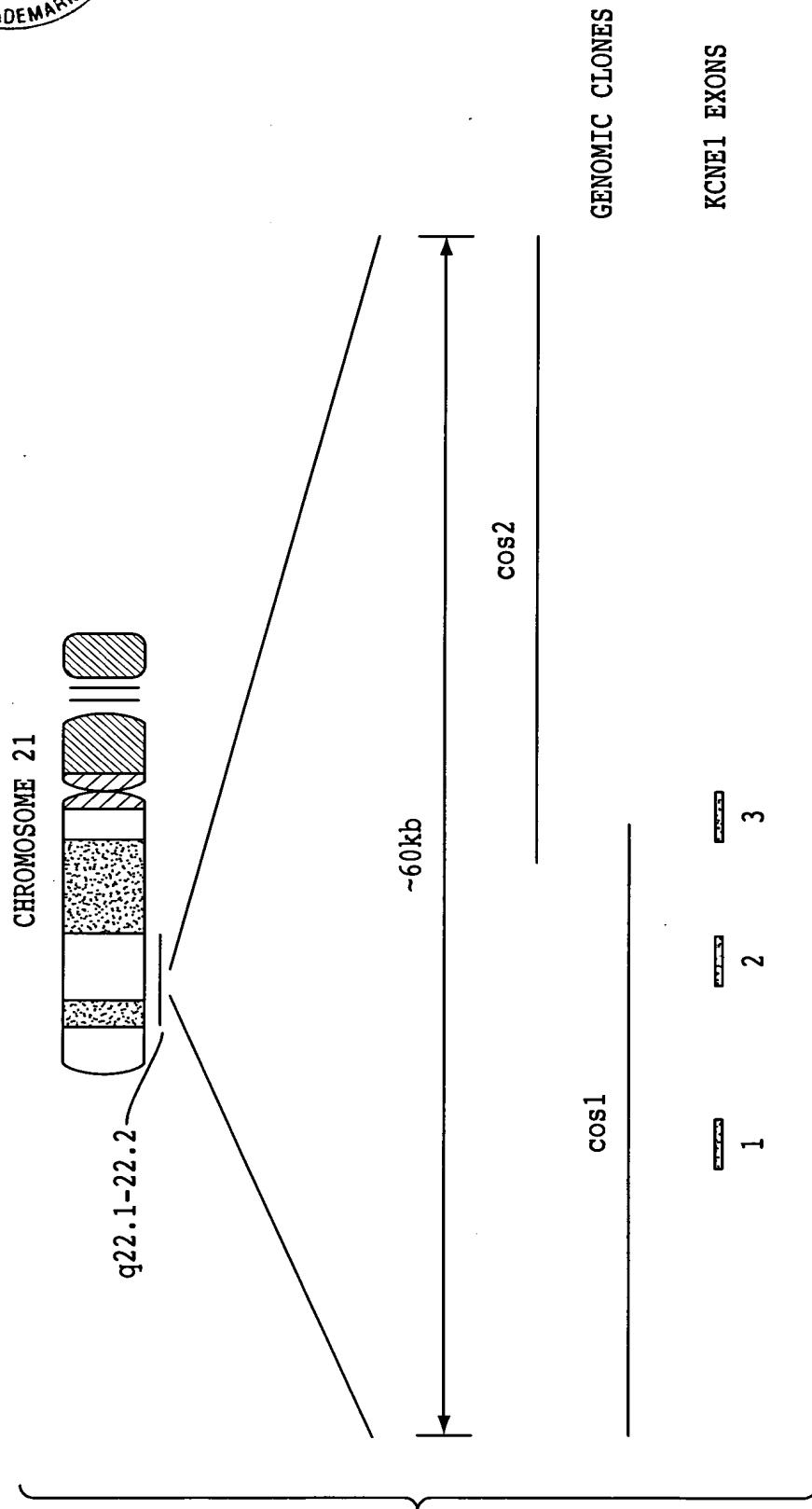


FIG. 17



KVLQT1 - A Long QT Syndrome Gene
Mark T. KEATING et al. - Atty Ref. 2323-151
SN 09/597,732, filed 19 June 2000

ACACCCGGCTCTCTGGCATCTCAGACCCGGAAAAATCCCTCTGCTTCTGGCCAGTTACACAATCATCAGGTGAG-81
CCGAGGATCCATTGGAGGAAGGCATTATCTGTATCCAGAGGAAATAGCCAAGGGATATTCAAGGGTGTGCCCTGGGAAGTTG-162
AGCTGCAGCAGTGGAACCTTAATGCCAGGATGATCCTGTCTAACACCCACAGCGGTGACGCCCTTCTGACCAAGCTGTGG-243
M I L S N T T A V T P F L T K L W -17
CAGGAGACAGTCAGCAGGGTGGCAACATGTCGGGCTGGCCAGGTCCCCCGCAGCGGTGACGCCAAGCTGGAGGCC-324
O E T V O O G G N M S G L A R R S P R S G D G K L E A -44
CTCTACGTCCCTATGGTACTGGGATTCTCGGCTTCTCACCCCTGGGCATCATGCTGAGCTACATCCGCTCAAGAACGCTG-405
L Y V L M V L G F F G F F T L G I M L S Y I R S K K L -71
GAGCACTCGAACGACCCATTCAACGTCTACATCGAGTCCGATGCCCTGGCAAGAGAACAGGCCTATGTCAGGCCGG-486
E H S N D P F N V Y I E S D A W Q E K D K A Y V Q A R -98
GTCCTGGAGAGCTACAGGTCGTGCTATGTCGTTGAAAACCATCTGGGCATAGAACACACACACCTCTGAGACG-567
V L E S Y R S C Y V V E N H L A I E Q P N T H L P E T -125
AAGCCTTCCCCATGAACCCACCACTGGCTAACTGGACACCTCTGCTGGNNNNAGATTTCTAATCACATTCTCA-648
K P S P * -1702
TACTCTTATTGTGATGGATACCACTGGATTCTTTGGCTGTTGTAANGGTGAGGGTGGATTAATGACACTGTTCA-729
CTGTTCTCTAAATCACGTTCTTGATAGACTGTCAGTGGTTCCCCATATCTGCCCTGCTAAATTAGCA-810
GAATCCCTGAGGACATGGCTCTGAGAATAGCAGCTGCATTCCAGACTCCCTTGCAAGCTAGCAAGGTTGTGACTAAG-891
CCCTGGCAGTAGGCATGGAAGTGAAGACTGTAATGTCAGTAATCTTGGAAAGAAAAGAACGTCGCCCTAACTAACCT-972
TGTCTGCTTCCCAGTGGCTGGATGTGGAGGGAGGTGGAGAGCAGTTATGAGACTGGAAAGTTCGGGGCACTCAAAGAGCC-1053
ACACACATCTGGGCTGGGCACGTGGATCCCTTACCAACCCACCCAGGGCAGATTACAGGAAGAGAAAATCCACTCCAC-1134
TCTTCCTTAAGGCCACTGTTATTCTGATCTGTTAAGGTCGAGAATCAATGCCCTTACTGATACACCTACCTTATAGGAC-1215
TGAACCTAAAGGCATGACATTCCATACTTGTCAACAGCACACACTGATCTGCCCTGTCACCTACCTGCTCACTCTG-1269
GGCTCTATCCTCCTGCCCTTCCGCTTCCACTCCTGCCATCCTGACACATCTCCCTGAAACACACAGG-1377
CACATACACTCATATAACATAGACACACATACACCTCAATCTAGAAAGAACCTGTTGTCAGGGCTGAGATGGAGGAG-1458
AAAAAAATGCCCTTCAGAATGCATACCAAGGGGAAGGTGCTCGGTCACTGTGGAGCAGGGAAAGGTGCCCTTCACTCCC-1539
CGAGAGCCAGGGGAAGGGAGTGGCTCTGGCAGAGAGGGACACATAGCACTGGGGTGGCAGGTCTTTGAGGTGATGGGCC-1620
GGTTTGAGATGAATTGTATCCCCAAAAGACAGGTACCTCAATGTGACCTAATTGGAAATAGACTTGCAGAT-1701
G(A)n -1702

FIG. 18